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CASE REPORT

Anterior clinoidal meningioma coincidental with bilateral intracranial aneurysms

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Abstract

Coexistence of aneurysms and brain tumors is a rare occurrence. Coincidence is highest in patients with meningiomas rather than other types of tumors. We report a case in which a meningioma of the left anterior clinoid process was coexisting with a right middle cerebral artery (MCA) and a left anterior cerebral artery (ACA) aneurysm. While the right MCA aneurysm was detected preoperatively, the left ACA aneurysm was not detectable, being concealed by the major finding of the region. This report focuses on pitfalls of diagnosis and questions the surgical planning in aneurysms concealed by coincidental brain tumors. Hippokratia 2011; 15 (4): 353-355

Key words: meningioma, intracranial aneurysm, anterior clinoid process

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There is a paucity of data regarding the true incidence of aneurysms in patients with brain tumors[1-3]. Reports indicate an incidence of approximately 0.5%[4]. The most frequently involved intracranial tumor associated with aneurysms is meningioma[1]. Endovascular techniques may be employed prior to tumor surgery, when the two coincidental pathologies are known. This case highlights the importance of considering underlying vascular abnormalities when resecting tumors, raising questions of surgical planning and pitfalls of diagnosis, which may limit awareness and influence treatment strategy in aneurysms concealed by coincidental brain tumors.

Case report

A 55-year-old female patient presented with two episodes of vertigo. Computer tomography (CT) revealed a hyperdense lesion in contact with the left anterior clinoid process, compatible with a meningioma. No intravenous contrast medium was administered at that time because of allergic history. Magnetic resonance imaging (MRI) showed a parasellar lesion with the charactestics of a meningioma (diameter ca. 3 cm) of the left anterior clinoid process, pressing the ipsilateral MCA anteriorly and pressing on the optic chiasm (Figure 1a).

The patient was referred to our institution for operative treatment. No focal neurological deficits were detected at the clinical examination. Immediate preoperative imaging with MRI and MRA revealed again an enhancing lesion at the left cavernous sinus region as well as a small aneurysm of the right MCA, which was only detected on retrospective review of the films.

The patient underwent a minimally invasive left supraorbital craniotomy and complete resection of the meningioma (Simpson II). Histology confirmed the presumptive diagnosis of meningioma. Postoperatively the patient developed a mild right hemiparesis, which improved with physiotherapy leaving no residual neurological deficit. A postoperative CT scan revealed a hemorrhage in the left sylvian fissure (Figure 1b). One week later, an MRI study showed no residual tumor, while no vascular lesion was reported.

The patient was closely followed clinically and radiologically thereafter. One year later MRI and MRA identified the known aneurysm of 6mm diameter at the bifurcation of the right MCA. A four-vessel digital subtraction angiography (DSA) revealed an aneurysm of the right MCA pointing anteriorly (Figure 2b) and a second small aneurysm of the left ACA pointing upwards (Figure 2c). This finding could presumably have developed after the resection. Careful reviewing of the preoperative neuroimaging revealed however a suspicious, obviously overlooked, finding on preoperative MRA (Figure 2a), thus emphasizing the value of a 4-vessel DSA.

The possibility of coiling was discussed, but neuroradiologists considered the aneurysm inappropriate for endovascular treatment. The patient underwent a right pterional craniotomy and clipping of the right MCA aneurysm (Figure 3a). Clipping of the left ACA aneurysm through this approach during the same operation was attempted, but not possible intraoperatively, due to the aneurysm direction and the suboptimal angle of approach. The post-operative course was uneventful and the patient was discharged home on the seventh post-operative day neurologically intact.

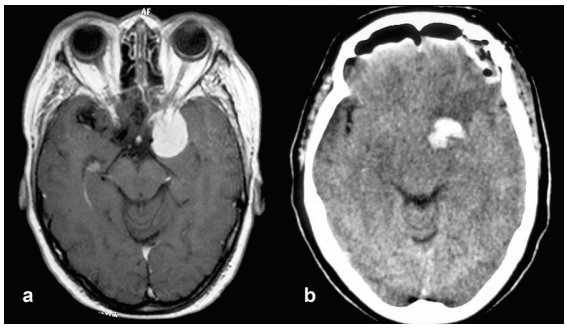

Figure 1 a.: Preoperative post gadolinium MRI T1w axial showing a meningioma of the left anterior clinoid, b. Post-operative CT scan showing hemorrhage at the resection site

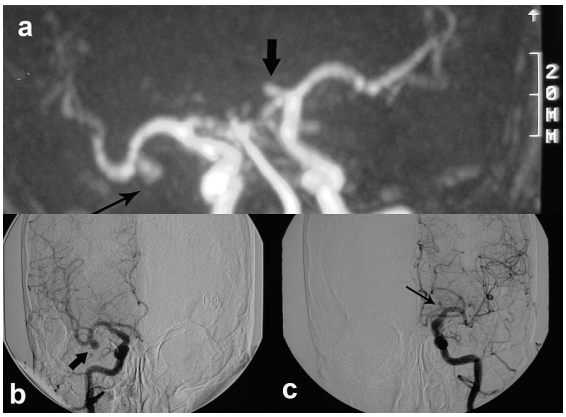

Figure 2 a.: MRA revealing a right sided MCA aneurysm (thin arrow) and a left sided ACA aneurysm (thick arrow), b. DSA showing the right sided MCA aneurysm (arrow), c. DSA showing the left sided ACA aneurysm (arrow)

The alternative of endovascular treatment was explained and offered to the patient for the contralateral ACA aneurysm. However, the patient, having developed a relationship of trust with her surgeon, chose to be operated. Hence, six months later a left pterional craniotomy and clipping of the left ACA aneurysm was performed (Figure 3b). Once again the post-operative course was uneventful and the patient was discharged without any neurological deficits.

Discussion

Coincidence of brain tumors and aneurysms is rare[5]. There is a paucity of data regarding the true incidence of aneurysms in patients with brain tumors[4]. Reports indicate an incidence of approximately 0.5%[4], which is estimated to be lower than the true incidence because four-vessel angiographic studies are not always performed in patients with intracranial tumors. Presenting symptoms in patients with coexisting brain tumors and aneurysms are tumor related in 70% of patients, aneurysm-related in 22%, and tumor- and aneurysm-related in 6%[6-7].

The most frequently involved intracranial tumor associated with aneurysms is meningioma[1,3]. Numerous reports of meningiomas coexisting with intracranial aneurysms have been published[1-3, 5, 7-9].

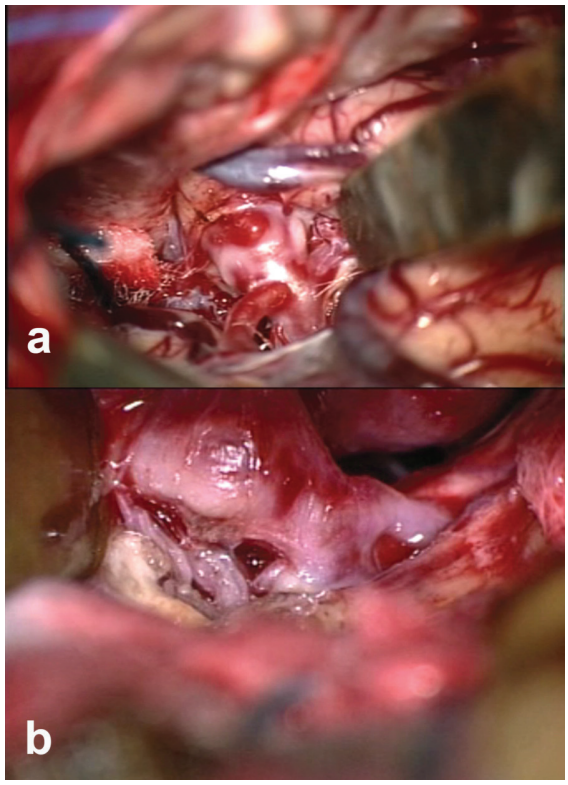

Figure 3 a. Intraoperative photo of the right-sided MCA aneurysm, b. Intraoperative photo of the left sided ACA aneurysm

Aneurysms are more often associated with skull base tumors in comparison to convexity tumors. There is a high frequency of convexity tumors with middle and anterior cerebral artery aneurysms, while basal meningiomas are more often associated with internal carotid artery and vertebrobasilar aneurysms[5].

Several speculative hypotheses have been proposed regarding the relationship between tumors and coexisting aneurysms[4]. One of the predisposing factors suggested that the development of an aneurysm (probably reasonable for highly vascular tumors such as meningiomas) is the increase in regional cerebral blood flow, since these aneurysms often seem to be related to the arteries that supply the tumors. In bilateral aneurysm cases however, such as in our patient, this hypothesis may explain the presence of the ipsilateral, but not the contralateral aneurysm. Bilateral middle cerebral artery aneurysms are rarely reported in literature[4].

The mortality of patients with both a brain tumor and aneurysm is as high as 38% [4]. Silent intracranial aneurysms distant from the lesion to be treated have usually little relevance to surgical management[4]. However, aneurysms adjacent to convexity and basal meningiomas are potentially an additional hazard in surgical treatment[4]. Careful stepwise procedures are essential to treat the aneurysm and the tumor simultaneously[2]. Clipping

the aneurysm safely after piecemeal removal of the tumor which is totally extirpated without fear of aneurysm rupture is a safe option[2]. Endovascular techniques may be employed prior to tumor surgery, when the two coincidental pathologies are known. The development and availability of endovascular treatment alternatives allows coiling of aneurysms preoperatively, thus making the surgical excision of an intracranial tumor co-existing with an incidental aneurysm safer[3, 10].

In our case, the conspicuous finding was obviously overlooked by both radiologists and surgeons and was only detected on retrospective review of the imaging. Such pitfalls limit the awareness and influence surgical planning, not allowing for endovascular preoperative treatment of the aneurysm. Furthermore, they emphasize the value of preoperative MRA or 4-vessel DSA even with the slightest degree of suspicion.

Conclusions

This case emphasizes the importance of considering underlying vascular abnormalities when resecting tumors. It also raises questions of surgical planning and pitfalls of diagnosis, highlighting the careful review of preoperative imaging and the value of preoperative angiography.

Conflict of Interest Statement

On behalf of all authors, it is hereby stated that there is no conflict of interest regarding this paper.

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CASE REPORT

Nephrogenic diabetes insipidus : Old deletion, new effect. A case report of a family from Greece

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Abstract

Congenital, X-linked, Nephrogenic Diabetes Insipidus (NDI) is a rare disorder in which the kidney is insensitive to the antidiuretic hormone, vasopressin. The gene responsible for this type of NDI, the V2 vasopressin receptor, has been cloned and mapped to Xq28.

We report the case of a boy, 2.5 month old, who presented with nephrogenic diabetes insipidus (NDI). The mother and the 7 year old sister of the boy also had the NDI phenotype but did not seek medical attention until the presentation of the boy to our department. The mutational analysis of the patient showed the R337stop mutation, also founded to the mother's genotype analysis. The allele separation in mother revealed the second X chromosomal allele with a 12-bp in-frame deletion. The same in-frame deletion was also found in his sister's genotype. This deletion of four amino acids (Arg-247 to Gly-250) has been previously described but was suggested not to be linked with the NDI phenotype. However, in our case, the only possible cause of NDI phenotype in the boy's sister was the 12-bp in-frame deletion. Hippokratia 2011; 15 (4): 356-357

Key words: nephrogenic diabetes insipidus, 12bp-ICL3 deletion mutant

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Nephrogenic diabetes insipidus (NDI) is a disease characterized by the inability of the kidneys to concentrate urine in response to vasopressin. It can be divided in two forms, acquired or congenital NDI. Acquired NDI is the most common form and can be seen in several pathological conditions such as renal hypoplasia, interstitial nephritis, chronic renal failure, obstructive uropathy, sickle cell disease, hypokalaemia, hypercalcemia. Congenital NDI is a rare disorder secondary to either mutations in the AVPR2 gene that codes for the vasopressin V2 receptor or to mutations in the AQP2 gene that codes for the vasopressin-depended water channel aquaporin 2. Mutations in the gene located on Xq28 coding for the V2 receptor of vasopressin (AVPR2 gene) are responsible for the X-linked mode of inheritance, while mutations of AQP2 gene are found in the autosomal recessive and dominant traits of the disease[1,2]. About 90% of symptomatic patients are males. Although most of the female carriers with heterozygote V2R gene mutations are usually asymptomatic, some female carriers have been reported with mild or even severe NDI phenotype due to skewed X-inactivation, as it is known till now[3]. The first manifestations can be recognized during the first weeks of life. Infants suffer from nonspecific symptoms such as polyuria, polydipsia, unexplained fever, vomiting, anorexia, failure to thrive. If this condition is not recognized early, congenital NDI children may experience several episodes of hypertonic dehydration that can be complicated by cerebral edema and convulsions.

Case report:

A 2.5 month old boy presented in the emergency department of our hospital with fever (38.5°C) twice a day and no other symptoms for the last two days.

On examination his weight and height were at 90th percentile for age, his blood pressure was 96/61 mmHg (on the 50th percentile for age, gender and height). From the clinical examination there were normal findings. Laboratory investigations revealed: sodium 165mmol/L, chloride 130mmol/L, serum osmolality 334mmol/L, urine osmolality 117mosm/kg, urine S.G. 1001. Sepsis control was negative. During his stay in hospital his urine volume was till 9ml/kg/h. ADH was 24ng/ml, aldosteron and renine between normal ranges.

Based on the hypernatriaemia, serum hyperosmolality, urine hyposmolality, and low SG of urine, a d DAVP (1-deamino-8-D-arginine vasopressin) test was performed and the urinary osmolality remained unchanged. MRIs of brain and pituitary gland were normal, renal ultrasonography presented normal kidneys with anterioposterior diameter of left renal pelvic 6mm. Voiding cystourethrography revealed bilateral vesicoureteral reflux (3rd grade on the right and 4th grade on the left side).

Nephrogenic diabetes incipidus was diagnosed and genotype tests for the patient, his mother and sister were performed at the Institute for Biochemistry, Faculty for Medicine, University of Leipzig, Germany

The boy's sister (9 year old) and mother (37 year old) presented with polyuria, polydipsia (mother has been